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A CRITICAL REVIEW OF THE SURGICAL TREATMENT OF INFANTILE FIBROUS DYSPLASIA

A. ANDRISANO, P. CALDERONI, G. MIGNANI, M. MANFRINI
(Bologna)

A critical review of the surgical treatment of 65 cases of infantile fibrous dysplasia demonstrated that «circumscribed» forms of the disease generally do not require any type of surgical treatment, while "extended" forms, as well as Albright's syndrome, require early surgical treatment aimed at preventing the development of skeletal deformities which are difficult to correct. Prophylactic intramedullary nailing with nails of suitable calibre most effectively achieves this goal.

During growth the localisation of fibrous dysplasia in the tubular bones, particularly in "extended" forms of the disease and in Albright's syndrome, is a condition which may lead to progressive skeletal deformity, often severe, as well as the occurrence of pathological fractures (Campanacci *et al.*, 1975; Harris *et al.*, 1962; Dohler and Hughes, 1986). In addition to sarcomatous degeneration, which is rare (Campanacci *et al.*, 1979), these mechanical complications constitute the principal indications for the treatment of areas of dysplasia which, at the end of growth, lose their potential to proliferate and "mature" into dense fibro-osseous tissue (Campanacci and Leonessa, 1970; Campanacci, 1981-86). Some authors (Dohler and Hughes, 1986; Stephenson *et al.*, 1987) have proposed surgical treatment as a means of preventing the mechanical complications. The purpose of this study is to critically review 65 cases of fibrous dysplasia submitted to surgical treatment, particularly in relation to the results obtained with the various methods used.

MATERIAL AND METHOD

The clinical and radiographic data of 65 patients (38 males and 27 females) aged on average 8.6 years (range 1 to 16 years) at the onset of treatment were analysed. The histological diagnosis and surgical treatment were both carried out at the Rizzoli Orthopaedic Institute between 1956 and 1986. Surgery involved two different skeletal sites in 3 patients. Overall, 68 localizations treated surgically were evaluated; the various sites involved are reported in Table 1. Thirty-one patients were operated on more than once (average 3 times), and a total of 124 operations were performed (Table 2), with an average follow-up of 6.6 years ranging from 6 to 27 months.

From: Divisione di Ortopedia e Traumatologia Pediatrica, Istituto Ortopedico Rizzoli, Bologna.

Table 1
SITE OF OPERATION

Pelvis	1
Femur	45
Tibia	16
Humerus	5
Forearm	1
Total	**68**

Table 2
OPERATIONS CARRIED OUT

Emptying and curettage	83
Rigid osteosynthesis	31
Intramedullary nailing	8
Resection	2
Total	**124**

The cases were divided into circumscribed and extended types of the disease. By circumscribed fibrous dysplasia we mean any lesion the radiographic extent of which is less than one-fourth of the bone segment involved, with involvement of only one cortex; by extended fibrous dysplasia we mean any lesion of greater extent, with involvement of both hemidiaphyses. In our cases, the indications for surgery differ considerably for the two types and the results were evaluated separately.

The surgical treatment of circumscribed forms of the disease (37 patients) was limited in 35 patients to incisional biopsy and curettage. In two cases involving the femoral diaphysis, diaphyseal hemiresection was performed.

In extended forms of the disease and in Albright's syndrome (84 sites operated on in 28 patients) the indications for surgery were based on the mechanical complications (deformity, pathological fractures, often simultaneous) with the object of correcting them or preventing progression. Many of these lesions required successive operations.

The surgical methods used were divided into three groups: emptying and curettage, osteosynthesis with plate and screws, and intramedullary osteosynthesis.

The result of each operation was judged to be positive or negative depending on whether it had determined healing of the dysplastic area or had prevented mechanical complications.

RESULTS

In circumscribed forms of the disease, emptying and curettage produced

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negative results in 60% of the cases. This high failure rate may, however, be attributed exclusively to the persistence of an osteolytic area; none of these cases resulted in pathological fractures or severe bony deformity at a later date (Table 3).

In extended forms of the disease and in Albright's syndrome (Table 4) emptying and curettage and osteosynthesis with plates and screws did not produce positive results (Fig. 1), whereas out of 8 cases treated with intramedullary fixation there were only two negative results (25%). In one of these patients there was progressive deformity after operation, the cause of which was attributed to inadequate fixation (Rush nail). The other failure was a case of femoral nailing according to the Küntscher method, carried out after five other operations had failed. There was postoperative infection and, after removal of the nail, severe limb length discrepancy.

Table 3
CIRCUMSCRIBED FORMS (37 PATIENTS)

Type of operation		Positive	Negative
Emptying and curettage	38	16 (42%)	22 (58%)
Resection	2	1 (50%)	1 (50%)

Table 4
EXTENDED FORMS OF THE DISEASE AND ALBRIGHT'S SYNDROME (28 PATIENTS)

Type of operation		Positive	Negative
Emptying and curettage	45	0	45 (100%)
Rigid osteosynthesis	31	0	31 (100%)
Intramedullary nailing	8	6 (75%)	2 (25%)

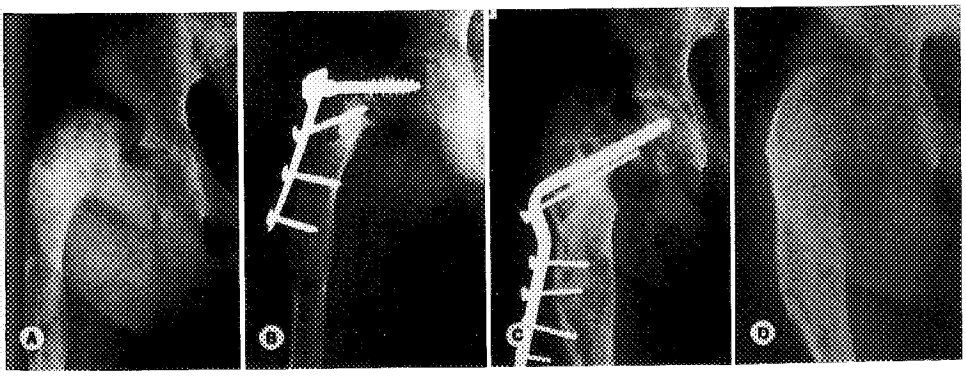

FIG. 1. - A) Patient aged 9 years; extended fibrous dysplasia with severe coxa vara previously operated on by three emptyings and curettage. B, C) Two osteotomies were carried out which resulted in loosening of the fixation. D) Two years after removal, the deformity progressed. Negative result.

DISCUSSION

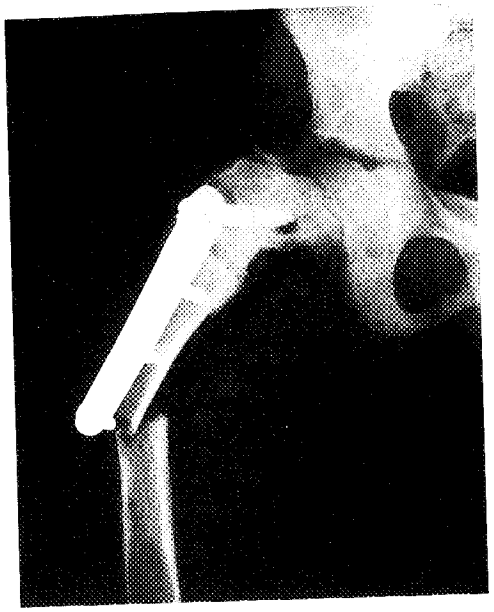

FIG. 2. - Fracture at the apex of the plate.

Our disappointing results affirm that in fibrous dysplasia during childhood, it is necessary, in the first place, to establish the extent of the dysplastic area, and whether it is circumscribed or extended.

In circumscribed fibrous dysplasia, given the minimal aggression of the lesion, it is sufficient to monitor progression, intervening surgically only if there are complications. When there are doubts about the diagnosis which necessitate biopsy, this must be conducted according to traditional protocol (Boriani *et al.*, 1984) proceeding with emptying and curettage only after the diagnosis has been confirmed.

In extended forms of the disease and in Albright's syndrome, on the contrary, it may be necessary to intervene early, before deformity occurs which is then extremely difficult to correct surgically. In these cases, emptying and curettage should be avoided; in addition to not producing healing of the osteolytic area, they do not in any way modify the aggressive progression of the disease.

The correction and prevention of deformity by rigid fixation with compression plates constantly failed in our cases and this was frequently associated with mobilisation of the plates, recurrence of the deformity, or fracture of the bone at the lower margin of the plate (Fig. 2) due to the poor holding quality of the pathological bone, or the concentration of tension in the bone plate interface.

Of the various methods used in this series, intramedullary nailing indubitably produced the best results (Fig. 3), thus confirming the experience previously reported by other authors (Seddon, 1967; Falcone and D'Arienzo, 1977; Connolly, 1977; La Terra and Ventura, 1985; Dohler and Hughes, 1986; Stephenson, 1987). In our opinion the relatively high percentage of negative results reported with this method may be prevented by using nails of a suitable calibre and by intervening early, before the patient has been submitted to multiple previous operations and before severe deformity occurs.

DISCUSSION

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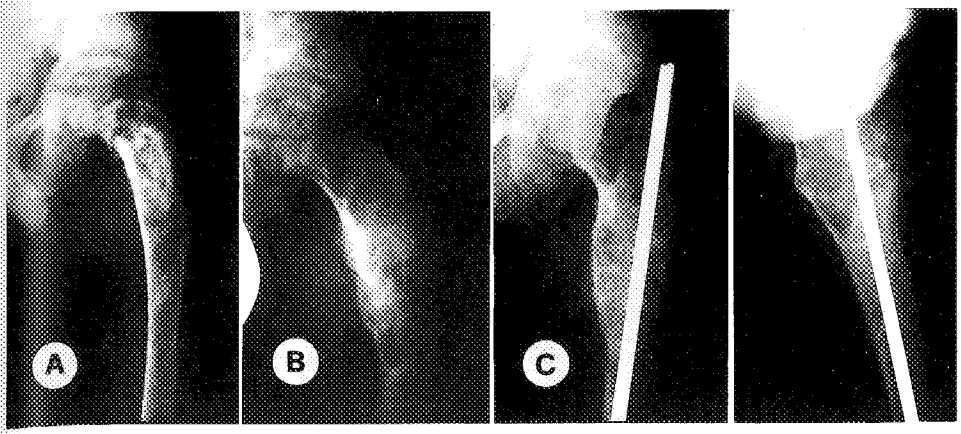

FIG. 3. - A) Example of extended fibrous dysplasia showing fracture and varus deformity. B) Valgizing osteotomy with rigid osteosynthesis was carried out, but because of the loosening of the plate and recurrence of the deformity, C) after 7 years, further corrective osteotomy and intramedullary nailing were carried out. The follow-up result after one year was positive.

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